

CAB19961

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## Product Information

<b>Product SKU:</b>	CAB19961	<b>Gene ID:</b>	1476	<b>Size:</b>	20uL, 100uL
<b>Clone No:</b>	-	<b>Host Species:</b>	Rabbit	<b>Reactivity:</b>	Human,Rat

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## Additional Information

<b>Observed MW:</b>	13kDa	<b>Conjugate:</b>	Unconjugated
<b>Calculated MW:</b>	11kDa	<b>Isotype:</b>	IgG

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## Immunogen Information

**Background:** The cystatin superfamily encompasses proteins that contain multiple cystatin-like sequences. Some of the members are active cysteine protease inhibitors, while others have lost or perhaps never acquired this inhibitory activity. There are three inhibitory families in the superfamily, including the type 1 cystatins (stefins), type 2 cystatins and kininogens. This gene encodes a stefin that functions as an intracellular thiol protease inhibitor. The protein is able to form a dimer stabilized by noncovalent forces, inhibiting papain and cathepsins I, h and b. The protein is thought to play a role in protecting against the proteases leaking from lysosomes. Evidence indicates that mutations in this gene are responsible for the primary defects in patients with progressive myoclonic epilepsy (EPM1). One type of mutation responsible for EPM1 is the expansion in the promoter region of this gene of a CCCCCGCCCCGCG repeat from 2-3 copies to 30-78 copies.

**Recommended Dilution:** WB,1:500 - 1:2000 IF/ICC,1:50 - 1:200

**Synonyms:** PME; ULD; CST6; EPM1; STFB; CPI-B; EPM1A; TB

**Purification Method:** Affinity purification

**Immunogen:** Recombinant fusion protein containing a sequence corresponding to amino acids 1-98 of human CSTB (NP\_000091.1).

**Storage:** Store at -20°C. Avoid freeze / thaw cycles. Buffer: PBS with 0.01% thimerosal, 50% glycerol, pH7.3.